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THE DEGENERATIVE DISEASES OF THE SPINAL CORD, WITH THE DESCRIPTION OF A NEW TYPE.[1]

By CHARLES L. DANA, A.M., M.D.

THE primary degenerative diseases of the spinal cord as now known are:

Locomotor ataxia.

Lateral sclerosis.

The combined scleroses, multiple sclerosis, peri-ependimal sclerosis.

The progressive muscular atrophies, including amyotrophic lateral sclerosis.

Neuroglia sclerosis.

These degenerations have all a certain degree of kinship. Their causes are in many respects the same, the course of all is uniformly progressive, and one not very unfrequently complicates another.

The sharpest distinctions are found between those affecting the gray matter and those affecting the white. Degenerative process affecting the former tissue are much rarer and their course is more rapid and fatal.

It is a curious fact that the anterior and central parts of the gray are almost exclusively affected, consequently we have clinically only progressive muscular atrophy and its congeners, bulbar paralysis, ophthalmoplegia externa and amyotrophic lateral sclerosis.

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[1] Read at a meeting of the N. Y. Neurological Society, Feb. 3, 1891.

The degenerations of the white matter are more common, slower in course, different in etiology and much more varied in symptomology. As a rule the gray matter is not much involved except secondarily and late.

The posterior roots and intervertebral ganglia are often diseased. Experiments seem to show that the anterior gray matter is supplied by a special plexus of vessels (*tractus arteriosus anterior* of Kudyi), while the lateral and posterior parts are supplied by other special plexuses (*tractus arteriori postero laterales*. It is further true that the gray matter is dependent on the blood supply for its integrity, while the health of the white matter depends largely on the nerve-cells to which its fibres are connected.

Among the various primary degenerations of the spinal cord, the combined scleroses are those concerning which our knowledge is most unsatisfactory.

1. We have one quite well established clinical and pathological type, that of Friedreich's ataxia.
2. We have another class in which the scleroses complicate paralytic dementia.
3. We have also the clinical type known as ataxic paraplegia.
4. There is, however, quite a number of cases in which combined sclerosis has been found after death, but in which the symptoms during life were of most irregular character. In 1887 I collected ("Medical Record," July 2, 1887), the histories of forty-five cases of combined sclerosis, with autopsies. Since then there have been reported thirteen other cases not including several of Friedreich's ataxia.[2]

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[2] Since the publication of my article on ataxic paraplegia in 1887, there have been reported four cases with autopsy by Dr. J. J. Putnam ; one case by Dr. M. Clark, Brain, 1889, Part LI.

Adamkiewicz, Wien. Med. Press, 1. Centr. f. Neur., 1888, p. 272.
Oppenheim, Centr. f. Neur., 1888, p. 649.
Glynn, Liverpool Medico-Chirurg. Journ., July, 1889.
Dr. Cane, Dublin Med. Journ., July, 1889. Dana, Brain, 1889, 1.
Whiton, Australasian Med. Gazette, April, 1889.
H. Brauen, Deut. Arch. f. Klin. Med., 1887-8, XLII., p. 459.
M. Sacelis, Reforma Med. Roma, 1888, IV., 1130, 11136.

Some of them presented during life the symptoms of ataxic paraplegia as described by Gowers, but others were of irregular course.

The question now arises, can we pick up any of these scattered ends and make of them anything definite and deserving a special name and type.

I believe that mainly through the efforts of Dr. J. J. Putnam, of Boston, this query may be answered in the affirmative. In the JOURNAL OF NERVOUS AND MENTAL DISEASE for February, 1891, he has described a group of cases of combined sclerosis of the spinal cord associated with diffuse collateral degeneration.

These cases clinically, etiologically and pathologically differ considerably from any of the other cases of combined sclerosis which I have seen reported.

In his description of one of them at the last meeting of the American Neurological Association, I was struck with its similarity to a case which had come under my observation At the time it occurred to me that we had here something peculiar, and my observations made independently of Putnam have, therefore, a special comparative value.

Dr. Putnam reports the history of four other fatal cases in which no post-mortem was made.

Among the cases heretofore reported by others, it seems to me that that of Sioli may be included as belonging to Putnam's group.

If this be so, we have six cases with autopsies and four without on which to base a description of the disease.

Such a description would briefly be somewhat as follows:

ETIOLOGY.

The patients were in most cases (7 out of 19) women. The ages ranged from 45 to 64 years. In some there was a neurotic inheritance, and in several lead was found in the urine at times. No distinct history of syphilis could be made out in any instance, nor were the patients alcoholic.

The disease ran a rather rapid course, varying from nine. months to four years, and averaging two years.

SYMPTOMS.

The symptoms began generally with numbness of the extremities, followed by progressive enfeeblement, and ending always in a preterminal paraplegia. Great emaciation and anæmia were present, and there was often an obstinate diarrhœa.

No paralysis of any special groups of muscles occurred until the final paraplegia set in.

There were in some cases anæsthesia and ataxia, but spastic symptoms with exaggerated knee-jerk and ankle clonus were the more common.

Lancinating or girdle pains were very rarely present.

The arms were affected, but less than the legs. The vision and other special senses and speech were not disturbed. Mental symptoms approaching dementia were observed in the terminal stages in some cases.

The general course was that of a rather rapidly progressive affection causing paræsthesia and sometimes anæsthesia of the extremities, especially the lower, with progressive weakness of the extremities. This was associated with anæmia, general muscular emaciation, diarrhœa, ending in a paraplegia.

PATHOLOGICAL ANATOMY.

The pathological appearances of the spinal cord in this group of cases is described by Putnam in words which so perfectly fit my own case that I cannot do better than use them.

"In all the cases, two sets of changes in the cord are recognizable, one of older date, consisting in a relatively dense sclerosis in the posterior columns and in the lateral column (mainly confined to the pyramid tracts); and one of subacute character, and evidently of quite recent occurrence. This sub-acute process was, as regards the white columns, partly in new tracts, partly around the borders of the more dense sclerosis, and was chiefly characterized by the perforated appearance, now familiar to every pathologist, which actually results, perhaps, from the post-mortem changes induced by hardening, but indicates a somewhat rapid destruction of nerve-tubes with the œdematous distention or destruction of the intervening septa, associated with the formation of granule cells.

In the gray horns, the degenerative change (partly recent, partly of older date) was indicated by a disintegration of nerve-cells. In certain parts of some of the cords, indeed, the almost entire destruction of nerve-cells shows that, in them, the ganglionic matter as well as the white columns had borne its full share of the brunt of the degenerative process."

The nerve-roots in certain regions, usually the posterior, lumbar or sacral, are somewhat involved. The peripheral nerves in the cases examined showed no especial change.

The brain and medulla are not affected.

The morbid process in the spinal cord is not primarily inflammatory, though some reactive meningitis may occur in the terminal stage.

The cases in question do not all resemble each other closely, but they do seem to be much more nearly allied to each other than to any known form of systemic spinal degeneration.

Their differentiation as a special group seems measurably justified by the following considerations :

Etiologically, it is peculiar in occurring so often in women and at a comparatively advanced period of life. Syphilis does not appear often to be a factor in its cause.

Symptomologically it is distinguished by its almost subacute course, by the presence of parasthesiæ and often of anæsthesia, with, as a rule, spastic symptoms, and finally paraplegia. Pain also is not common. Ataxic paraplegia, on the other hand, runs a very chronic course, anæsthesia is quite rare and paraplegia comes late.

Anatomically it is peculiar in the fact that the degenerative process is rapid, and accompanied with a peculiar collateral or terminal softening. The lesion is also less diffuse than in ataxic paraplegia, and involves the root-zones of the posterior column and the posterior columns generally, more than in the allied disease.

Its distinction from locomotor ataxia with secondary lateral sclerosis is in every way most marked.

It may be said, perhaps, to bear some resemblance to those secondary changes in the cord which occur in general paralysis. But none of the reported cases had this disease, though one was somewhat demented a short time before death.

The history of my own case, for the details of which I am indebted to Dr. Leo Etinger, house physician at the Montefiore Home, is as follows:

Babette S., 44, Prussia, wid.

Family history: Mother died at 80, suddenly, with symptoms of acute dyspnœa; father at 70, after losing his fortune, brooded over it, became somewhat demented. Three brothers alive and well, two sisters also; none dead. Husband died four years ago of some pulmonary trouble. His children by another wife, quite well. Thinks she had an aunt who may have had some paralytic trouble.

Personal history: Ill since July, 1889. Had measles as a child; was a fairly healthy, but delicate child. Went to school; found work easy; left at 14; married at 32. Menses at 12, were quite regular. Still has her menses, latterly more frequent; more blood lately; no miscarriages; no children. Eight years ago, while engaged with a thief, covered by the latter with the boxes and show-cases in the cigar-store owned by her husband. Later struck senseless by a blow in the face; remained in bed two weeks; then obliged to appear in court. Ever since her nerves have been unstrung. For the last three summers subject to diarrhœa. Her right leg had been swollen for some two or three years, but could do her work without trouble.

Last July (1889) attacked by weakness, vomiting, diarrhœa, and she believes some fever. Confined to bed for a week or ten days. Then got up. Returned to bed shortly; kept on going to bed and getting up for two months. In the beginning of September had some family trouble, which brought on an attack of screaming, crying; was entirely conscious. Same evening felt weak all over and had diarrhœa. Says that latter occurs after every excitement. Went to bed the same evening ; remained there ever since. Has had jerking movements of her legs, especially the right. Gradually lost her power in both legs, especially the left, which was always thinner. From time to time has a sensation of pins and needles in her heels. Has not had any pain, except with every change of weather she has queer, indescribable, somewhat painful feeling in both lower extremities, more marked in the right and in the thighs than in the legs. Finds that sometimes she passes too much water and at other times not enough. Two weeks ago was twice catheterized, not having passed her water for twenty-four hours. With passage of water sometimes had a movement of her bowels in spite of herself. Appetite is poor, sleep poor and restless. Sometimes the bowels do not move at all, at other times too much, with tenesmus.

Status præsens: A woman of slight build, skeleton small, eyes dark, height below medium. Mucus membranes very anæmic.

The patient on admission could barely walk with assistance. She was put in bed and her paraplegia speedily increased, but never became so great that she could not move her legs somewhat.

The legs and thighs were considerably wasted, the left more than the right. They were slightly drawn up and she suffered at times from painful contractions in them.

The knee-jerks and superficial reflexes were abolished. There was no cutaneous anæsthesia ; the presence of ataxia could not be satisfactorily tested; there was no pain other than that referred to. She had some disturbance of the bladder. Her bowels were usually constipated, but at times under nervous excitement she had an obstinate diarrhœa. The arms were not much affected, though they were weak.

She was at times delirious and had delusions.

Vision was not affected.

She gradually grew weaker, and six weeks after admission died.

At the autopsy only the brain and cord were examined. The brain appeared normal.

The dura mater of the spinal canal was normal. On opening it an extensive softening was apparent in the lower dorsal region.

Aside from these local disturbances here, there were no gross evidences of disease. On sections an apparent sclerotic change was observed in the lateral and posterior columns throughout the cord.

The specimens were hardened in Müller's fluid and stained with hæmotoxylon, aniline blue, and by Weigert's and other methods.

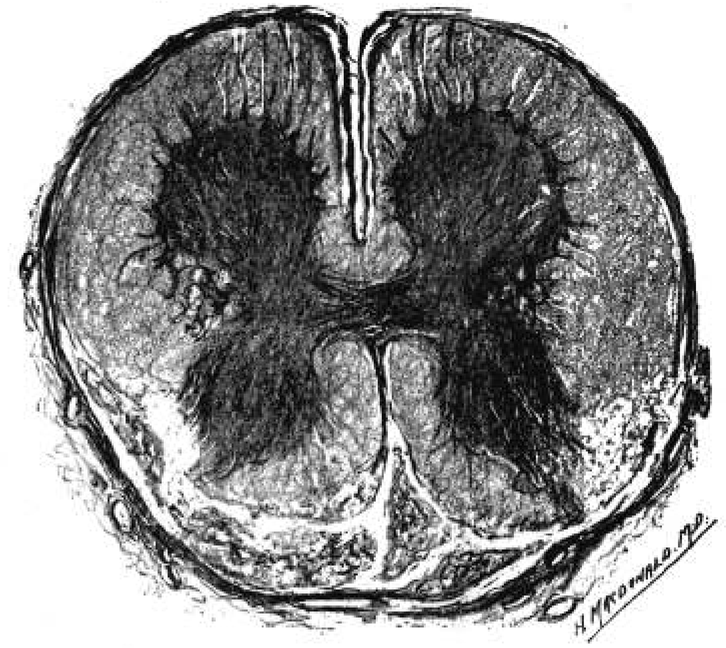

FIG. I.—Section at level of upper lumbar cord, showing the lateral and posterior degeneration and the terminal softening.

Microscopic examination showed the most active and recent process to be in the lower dorsal region posteriorly. Here the cord is softened by a process necrotic in nature, which is confined externally by the thickened meninges and internally by a pretty sharply defined wall of healthy cord tissue. Lower down, at the tenth dorsal, this process has extended further anteriorly, until it cut the cord nearly in half.

The softened area in some sections is filled with débris. of nerve tissue. Enlarged vessels are seen near the edge.

Outside the meninges is an exudation of lymph.

Besides this there was a degenerative sclerosis of the lateral and posterior columns throughout the spinal cord The process in the lateral columns was confined chiefly to the pyramidal tracts, but in the cervical region extended forward and involved the cerebellar and ascending lateral tracts. The posterior column shows a slight focus of beginning softening in the lower cervical region, and in the lateral columns the degenerative process seems rather subacute. It is not a hard selerosis. The spinal nerve roots are not affected except the posterior roots in the lumbar region.

The morbid process on the whole was apparently a sub-acute systemic degeneration, with a terminal focal softening. Associated with the last stage was a local reactive inflammation with some lymph exudation.

DESCRIPTION IN DETAIL.

On a close study of the changes of different levels one sees that at the *cervical level* the pia mater is somewhat thickened and shows evidence of a recent, but slight, meningitis, especially anteriorly and in the median fissure.

There are areas of sclerosis in the lateral and posterior columns in the regions indicated in the cuts.

The area in the lateral columns occupies the region of the crossed Pyr-tract, direct cerebellar and anterior ascending tracts. The lateral areas are studded with vacant spaces made by the swelling of the myelin sheath. These are more marked about the periphery of the diseased tracts. The process is more advanced on the left side. On each side there is a good deal of hyperæmia of the affected areas.

The process in the posterior columns is more advanced, than in the lateral and also attended with more hyperæmia, and occasional small hæmorrhages.

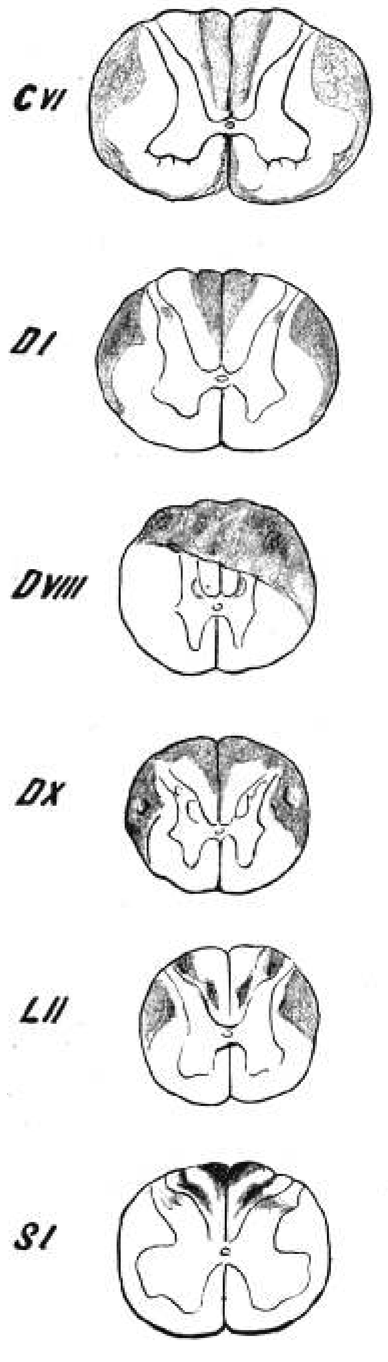

Fig. II.—Showing the affected area at different levels

Fig. III.—Showing the vertical distribution of the terminal softening.

The anterior median columns are slightly affected with the vacuolizing process, but show no softening.

There is a good deal of congestion of the central gray and anterior horns.

The anterior cervical-cells and nerve-roots are not especially affected.

Sections made through the upper part of *the dorsal cord* show similar changes to those above described.

Lower down the cord was too soft to cut, but the posterior and lateral sclerosis can be distinctly followed with the naked eye.

At about the level of the sixth dorsal, the posterior and lateral change begins to run together until about the ninth, cord is nearly cut in two by a disintegrating process.

At this neighborhood there is a reactive meningitis, with much exudation of lymph, but no pus.

At the level of the twelfth dorsal nerves, good sections can be obtained. These show a very distinct lateral sclerosis, occupying chiefly the region of the lateral tracts, and a posterior degeneration more marked on one side.

A destructive process lying within the pia has affected the posterior border of the posterior and lateral columns• There is here a very little of the vacuolization in the lateral and anterior median columns. The gray matter is congested. The cells of the column of Clarke are not affected except slightly on one side. The anterior horn cells are also but slightly involved. The root shows no degenerative changes.

At the *level of the first lumbar* there is decided degenerative change in the posterior columns, but very slight in the lateral.

There are a few cells of the column of Clark. The anterior and interior groups of cells of the anterior horns are atrophied.

There is a decided degenerative of the posterior roots, but not of the anterior.

The same description applied to the *level of the sacral.*

GENERAL REMARKS.

The *vessels of the cord* show no marked degree of degeneration, and the morbid processes can not be ascribed to their blocking up and obliteration. It seems to be rather a primary and rapid systemic degeneration, which was cut short by the focal softening that caused paraplegia and death.

The local process could not have caused the degenerations secondarily, as may be seen by its position, and the length of the history and symptoms point to the lateral columns as having been first affected.

The peculiar vacuolization of the degenerated areas, which was marked and characteristic in Putnam's cases, is present here, but not to any striking extent.

This process cannot be considered as indicating anything more than a rather rapid degeneration. It is seen in greater or less degree in almost all degenerative and inflammatory disorders, and is an œdematous swelling of the nerve-tubes which precedes their disintegration. Hence it is more marked in my case in the periphery of the degenerated areas, when the destructive process had not gone far.

The condition in which this vacuolization is still the prominent change in the system disease occurred in one case of ataxic paraplegia described by Dr. Græme M. Hammond, and reported by me in my article on this subject ("Medical Record," *loc. cit.*). In this case the patient was syphilitic and phthisical. He had marked ataxic and spastic symptoms, but the vacuolization and most of the morbid change was in the posterior columns of the spinal cord.

Pathologically, then, the principal characteristic of this case is its rapid course, its primary character, its systemic distribution, the terminal softening.

The brain, it should be added, was carefully examined. It hardened well and showed no evidence of morbid change in the meninges or exsection.